NMO antibodies should be ordered on all cases with acute optic neuritis

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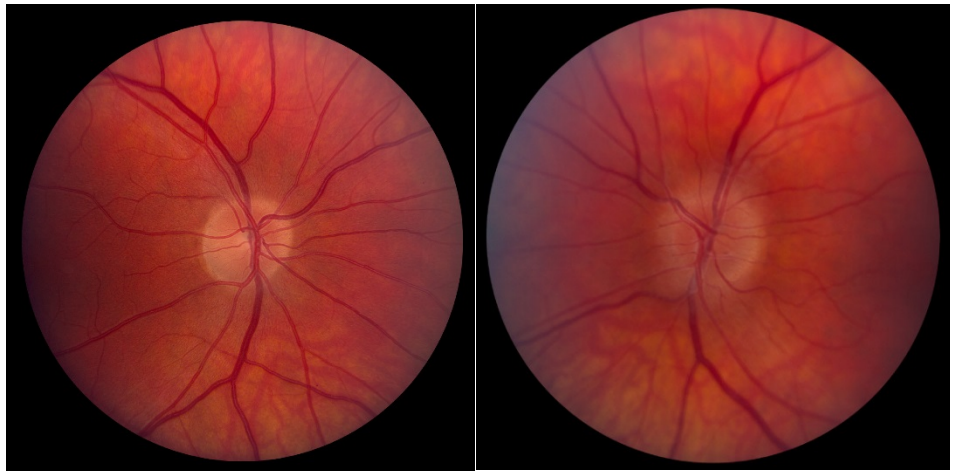
Two similar patients at presentation

- Two 32 yo white women
- No past medical history. No medication
- Pain around left eye for 1 week, worse with eye movements
- Blurry vision left eye 3 days ago, rapidly progressive since

- VA: 20/20 OD, Count fingers OS
- Color: 14/14 OD, no control plate OS
- Dense left RAPD
- Mild disc edema OS, normal retina
- Central scotoma OS, full VF OD



Left inflammatory optic neuritis

- No systemic symptoms, no fever, no neurologic symptoms
- MRI brain and orbits with contrast:
  - Left optic nerve enhancement
  - Two white matter lesions on brain MRI
- Syphilis negative
- [Lumbar puncture: 2 cells, normal protein, no oligoclonal bands]

- Receives 3 days of 1 g intravenous methylprednisolone followed by oral taper over 10 days
- Appointment with Neurologist to discuss treatment for multiple sclerosis versus observation

“Isolated inflammatory optic neuritis”

2 months later	
Patient 1	**Patient 2**
Much improved	Not improved
VA 20/20 OD, 20/25- OS Color 14/14 OD, 8/14 OS Small left RAPD Mild ON pallor	VA 20/20 OD, HM OS Color 14/14 OD, no control OS Dense left RAPD Diffuse ON pallor
Remained well without neurologic problems	Left eye never improved Optic neuritis OD, 3 months later Positive NMO antibodies

If I had known earlier it was NMO, I would have treated patient #2 differently

- I would have:
  - Given 5 days of IV steroids instead of 3 days
  - Considered plasmaphereses if vision did not improve after 5 days of IV steroids
  - Would have obtained a spine MRI urgently
  - Would have sent to Neurology urgently for treatment to prevent second episode (different treatments than those given for MS)

The only way to know that an optic neuritis is associated with NMO or MOG antibodies is to systematically test these antibodies in all patients with optic neuritis

Neuromyelitis Optica: Review and Utility of Testing Aquaporin-4 Antibody in Typical Optic Neuritis

Meagan Seay, DO, and Janet C. Rucker MD*†*

Abstract: Neuromyelitis optica (NMO) is an autoimmune, inflammatory demyelinating disorder often leading to severe vision impairment and disability. The discovery of a diagnostic biomarker, the aquaporin-4 antibody (AQP4-IgG), transformed the clinical diagnosis and treatment of NMO and broadened the spectrum of disease [NMO spectrum disorders (NMOSD)]. Though the antibody is highly sensitive and specific to NMOSD, routine testing in patients with typical optic neuritis is considered controversial. This article will provide a brief review of NMOSD and highlight the pros and cons of routine testing in typical optic neuritis.

Key Words: NMO, aquaporin-4 immunoglobulin, optic neuritis

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of an NMO patient and delivered to rats. These antibodies were found to bind AQP4 and to induce similar pathological changes seen in NMO.[13] Thus, identification of AQP4-IgG and its pathogenicity distinguish NMO and MS as separate entities.

The discovery of the AQP4 antibody precipitated widespread clinical testing, which led to broadening of phenotypes observed in NMO and new terminology: NMO spectrum disorder (NMOSD). In 2006, the revised NMO diagnostic criteria were changed to allow unilateral optic neuritis, though still required a history of both optic neuritis and myelitis.[14] Two of 3 supportive criteria were necessary: longitudinally extensive spinal cord lesion of at least 3 vertebral segments, magnetic resonance imaging (MRI) of the brain at onset that did not meet criteria for MS, and seropositive AQP4-IgG. In 2015, the criteria were revised once more, greatly expanding the clinical phenotypes.[15] Six core clini-

Aquaporin-4 antibodies

- Allow diagnosis of NMO-related disorder in a patient with one episode of isolated optic neuritis
- AQP4 antibody is specific for NMOSD
- Up to 40% of patients with atypical optic neuritis have positive AQP4 antibodies (bilateral simultaneous or rapidly sequential optic neuritis; unilateral painless optic neuritis; severe optic disc swelling; no recovery of vision)
- Up to 5% of isolated optic neuritis patients have positive AQP4 antibodies

Knowing results of AQP4 antibodies helps

- Treatment of NMOSD different from MS
  - Acute treatment is different
  - Follow-up is different
  - Some MS treatments (interferons, natalizumab, fingolimod) are harmful to NMOSD patients

In my clinic

- I routinely test patients with acute optic neuritis for AQP4 antibodies (and MOG antibodies too)
- I do not waste resources because I do not obtain tests that are not useful (I do not obtain “systematic tests”, such as RPR, ACE, ANA, Lyme, B12, etc….)